

Secundum atrial septal defect associated with a cleft mitral valve¹

Daniel J. Goodman, and E. William Hancock

From Cardiology Division, Stanford University School of Medicine, Stanford, California, U.S.A.

Five cases of atrial septal defect are reported, each of which was associated with a cleft in the anterior or posterior (or both) leaflet of the mitral valve. Only 3 had a characteristic murmur of mitral regurgitation, and 2 of these patients were thought to have a partial endocardial cushion defect. In two patients, the discovery of the mitral valve cleft came only at the time of operation, when a thrill was noted over the posterior left atrium. A high index of suspicion is necessary at the time of operation, because preoperative diagnosis may be extremely difficult. Closure of the atrial septal defect alone may materially worsen the effect of the mitral regurgitation. It is emphasized that the diagnosis of this type of mitral valve pathology in patients with an atrial septal defect is an indication for bacterial endocarditis prophylaxis.

A cleft in the anterior leaflet of the mitral valve is usually an integral part of the complex of lesions comprising an endocardial cushion defect (Weyn *et al.*, 1965). The partial form of this defect involves a low-lying atrial septal defect associated with a cleft in the septal leaflet of the mitral valve, and occasionally a similar cleft in the septal leaflet of the tricuspid valve. Four cases have been reported in the English language journals in which a cleft anterior mitral valve leaflet has been associated with a secundum type of atrial septal defect alone (Salomon, Aygen, and Levy, 1970; Pifarré *et al.*, 1968; Billig *et al.*, 1968; Edwards and Burchell, 1958). In 2 of the 3 cases diagnosed *ante mortem*, an endocardial cushion defect was suspected preoperatively (Pifarré *et al.*, 1968; Billig *et al.*, 1968).

We report here 4 cases of secundum atrial septal defect and 1 case of sinus venosus type atrial septal defect, each associated with a cleft in the mitral valve, 2 in the anterior leaflet, and 3 in the posterior leaflet. The cleft in each case rendered the mitral valve incompetent. Emphasis is placed on the varied presentation of this complex, its confusion with true endocardial cushion defect, and the importance of recognizing the existence of the mitral valve cleft. This last point is important from both a surgical and medical therapeutic point of view.

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Case material

Of a total of approximately 400 patients undergoing secundum atrial septal defect repairs performed in the years 1960 to 1972, 5 patients, ranging in age from 18 to 47 years, were discovered at operation to have a secundum type of atrial septal defect associated with a functionally important cleft in the mitral valve. All patients had a history taken, were examined, and had an electrocardiogram, chest x-ray, and cardiac catheterization before operation. Four patients also had a phonocardiogram. All patients were re-evaluated at from 18 to 78 months, mean 36 months, postoperatively. At this time, history, physical examination, electrocardiogram, chest x-ray (3 patients), phonocardiogram (4 patients), and cardiac catheterization (2 patients) were performed. The history and physical examinations, both pre- and postoperatively, and the surgical findings are summarized in the case reports. The results of electrocardiography and cardiac catheterization are presented in Tables 1 and 2. Phonocardiography was helpful in evaluating the nature and width of the split second sound and confirmed the findings noted on auscultation.

The preoperative chest x-ray consistently showed the typical changes expected in a patient with an atrial septal defect, including plethora in the lung fields and prominent main pulmonary arteries. Only Case 4, who had had mitral valve involvement by bacterial endocarditis, had an enlarged left atrium and left ventricle. Postoperatively, chest x-rays were available in Cases 1, 2, and 3, and in each case signs of plethora were no longer present. In addition, the heart size, though not grossly enlarged preoperatively, appeared smaller on the postoperative examination.

TABLE 1 *Electrocardiographic findings: preoperative and postoperative studies*

Case No.	P wave	QRS axis (degrees)	QRS duration (sec)	Morphology in V ₁ $\left[\frac{R:R'}{S} \text{ mm} \right]$
1	LAA	0	0.10	$\frac{2:4}{5}$
	LAA	0	0.10	$\frac{2:2}{6}$
2	LAA	0	0.09	$\frac{-:5}{2}$
	↓ LAA	-35	0.09	$\frac{1:6}{9}$
3	nl	+110	0.09	$\frac{11:-}{2}$
	nl	+75	0.08	$\frac{3:-}{-}$
4	LAA	+110	0.08	$\frac{1:1}{1}$
	↓ LAA	+110	0.09	$\frac{1:1}{2}$
5	LAA	Ind.	0.11	$\frac{1:7}{5}$
	nl	Ind.	0.11	$\frac{1:3}{7}$

Note: For each case, the preoperative electrocardiographic results appear above the postoperative study. LAA = left atrial abnormality; Ind. = indeterminate axis; ↓ = decrease; morphology of V₁ shows initial R wave and R prime wave over the S wave. If any deflection was not present, a dash replaces it in the Table. nl = normal.

TABLE 2 *Results of cardiac catheterization*

Case No.	RA	PAW/LA	PA	PVR	QP/QS
1	9/6/5	8/7/6	29/11/20	< 1.0	3.8
	3/3/2	10/8/7	14/6/9	0.5	1.0
2	6/5/3	8/7/5	29/8/17	1.0	3.2
	—	—	—	—	—
3	7/7/4	6/7/4	22/5/13	0.35	5.0
4	14/19/14	18/33/18	56/20/31	0.8	5.4
	—	—	—	—	—
5	13/5/8	13/5/8	82/30/46	12.0	2.0
	6/8/5	13/16/10	48/20/28	3.5	1.0

Note: The lower set of data for Cases 1 and 5 represent the results of the postoperative catheterization. The other patients had no postoperative study. RA = right atrial pressure a wave/v wave/mean; PAW/LA = pulmonary artery wedge or left atrial pressure a wave/v wave/mean; PA = pulmonary artery pressure systolic/diastolic/mean; PVR = pulmonary vascular resistance expressed in resistance units; QP/QS = ratio of pulmonary to systemic flow.

Case reports

Case 1

A 47-year-old asymptomatic man was seen in the cardiac clinic because of a heart murmur discovered during a routine check-up. Cardiac examination revealed a widely-split, fixed second sound with a loud pulmonary component a grade 2/6 pulmonary ejection murmur, and a grade 3/6 pansystolic apical murmur radiating into the axilla.

At operation, a 3.5 × 2 cm secundum atrial septal

defect was discovered. Examination of the mitral valve showed a 1 cm cleft in the anterior leaflet and a smaller cleft in the posterior leaflet. The posterior leaflet was noted to prolapse into the left atrium. The degree of mitral regurgitation was in agreement with that demonstrated preoperatively by angiography (grade 2/4). The cleft in the anterior leaflet alone was repaired, and the atrial septal defect was closed by primary suture. A residual thrill was noted at the completion of the operation.

At follow-up examination 26 months after operation,

the patient was asymptomatic. Cardiac examination showed a grade 3/6 pansystolic murmur of mitral regurgitation.

Case 2

An asymptomatic, 34-year-old woman was referred to Stanford University Hospital for evaluation of a heart murmur. Examination of the heart revealed a right ventricular impulse, a widely-split and fixed second sound with pulmonary components increased in intensity, an ejection click in the pulmonary area, and a grade 2/6 systolic ejection murmur in the pulmonary area. In addition, a grade 2/6 blowing systolic murmur was present at the apex, which radiated into the axilla.

At operation, a 3.5 × 2 cm ostium secundum atrial septal defect was found. Because of the preoperative suspicion of mitral regurgitation, the mitral valve was palpated through the atrial septal defect and felt to be normal. The septal defect was closed by primary suture, and the patient taken off bypass. At this time a thrill was felt in the posterior wall of the left atrium. The patient was replaced on bypass and the mitral valve was explored by direct vision. A cleft was found in the mid-portion of the posterior leaflet, and this was closed by suture. A very small residual thrill was present at the end of the procedure.

Postoperatively, the patient has remained asymptomatic. At a return visit 33 months after operation, physical findings included completely normal heart sounds and a grade 2/6 apical systolic murmur radiating into the axilla.

Case 3

An 18-year-old asymptomatic girl was referred to the cardiology clinic for evaluation of a heart murmur detected on a routine examination. Physical examination of the heart revealed a right ventricular impulse, a prominent ejection click in the pulmonary area, a widely-split and fixed second sound with an increased pulmonary component and a grade 2/6 systolic ejection murmur in the pulmonary area.

At operation, a typical 2 × 4 cm secundum type of atrial septal defect was found, and repaired by primary closure. After the patient was taken off bypass, a thrill was appreciated over the left atrium. The patient was replaced on bypass, and exploration of the mitral valve revealed a moderate-sized cleft in the posterior leaflet, which was repaired by suture.

Since operation the patient has been asymptomatic. At a follow-up visit 30 months later examination of the heart revealed no murmurs.

Case 4

This 31-year-old woman was referred for evaluation because of progressive congestive heart failure. Five years before, she had been treated successfully for subacute bacterial endocarditis, and had had symptoms and signs of mild congestive heart failure for three years, with progressively increasing symptoms in the three months before evaluation.

Cardiac examination revealed both right and left ventricular impulses, a fixed second sound with a loud pulmonary component, third and fourth gallop sounds, and

a grade 3/6 pansystolic murmur at the apex radiating into the axilla.

At operation, a sinus venosus type of atrial septal defect was present. In addition, a moderate sized cleft was found in the posterior leaflet of the mitral valve. Areas of this leaflet were seen to be involved by old endocarditis, and there were ruptured chordae to the posterior leaflet. The mitral valve was replaced with a Starr-Edwards prosthesis, and the atrial septal defect was closed by direct suture.

Postoperatively the patient has done very well, but has been bothered by recurrent episodes of atrial fibrillation necessitating cardioversion. Cardiac examination at follow-up visit 78 months postoperatively revealed normal prosthetic valve sounds and no murmurs.

Case 5

A 37-year-old woman was referred to Stanford University Hospital for surgical closure of an atrial septal defect and bypass graft for coronary arterial disease. She had a long, complicated history including chronic lung disease, diabetes, hyperlipoproteinaemia, hypertension, and severe angina, and suffered from marked dyspnoea on exertion. Cardiac examination revealed a widely split, fixed second sound, with a loud pulmonary component, a fourth heart sound, and a grade 2/6 pulmonary systolic ejection murmur.

At operation, a 4 × 6 cm fenestrated secundum atrial septal defect was found. Before bypass, a thrill was noted over the left atrium, and exploration of the mitral valve revealed a moderately large cleft in the anterior leaflet. Slight mitral regurgitation was present, too small to be demonstrated by preoperative ventriculography. The cleft was closed by suture, the atrial septal defect repaired by pericardial patch, and a coronary artery vein bypass graft to the right coronary artery was inserted.

Since operation the patient has had a pronounced decrease in the severity of both her angina and her dyspnoea on exertion, going from a class 3 to class 2 status (New York Heart Association classification). Cardiac examination 18 months after operation was unremarkable, except for a somewhat loud pulmonary component of the second sound.

Discussion

A cleft in the anterior leaflet of the mitral valve is commonly found in association with an ostium primum type of atrial septal defect (Weyn *et al.*, 1965). Rarely, an isolated cleft in this position has been reported to occur as a form of endocardial cushion defect (Edwards *et al.*, 1965). Only 3 antemortem cases have been reported in the English published material in which a cleft mitral leaflet has occurred with an ostium secundum type of atrial defect, and in each case the cleft was located in the anterior leaflet (Salomon *et al.*, 1970; Pifarré *et al.*, 1968; Billig *et al.*, 1968). In 2 of these cases, a loud murmur of mitral regurgitation was present, which suggested a diagnosis of endocardial cushion defect preoperatively.

In the 5 cases presented here, the correct diag-

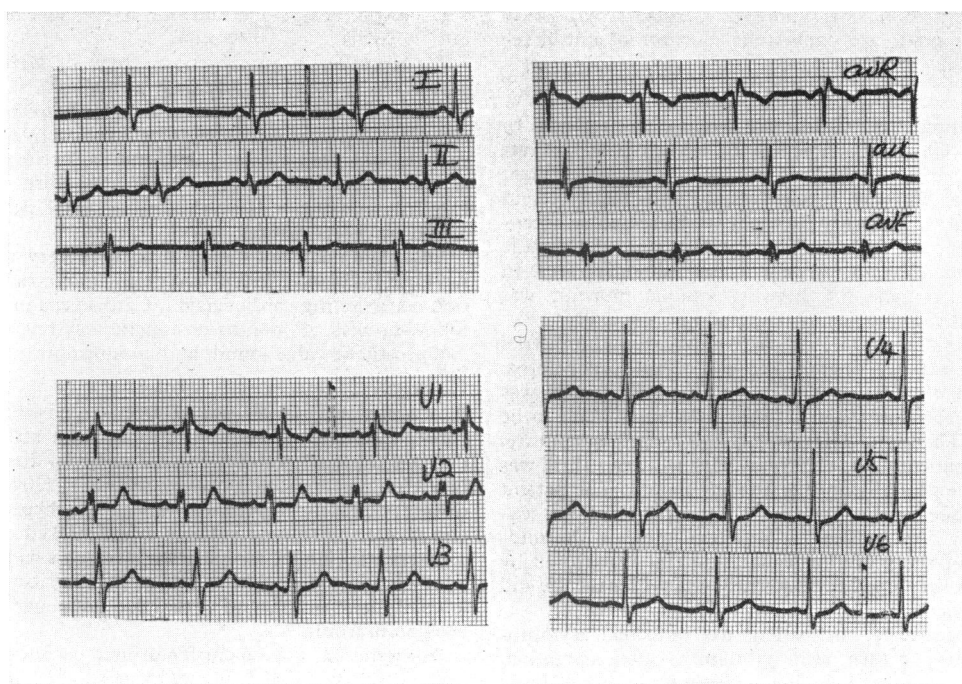


FIG. Preoperative electrocardiogram of Case 1, showing a frontal plane axis of 0° . The patient is in a sinus rhythm, there is a prominent late negative component of the P wave in lead V1, suggesting left atrial abnormality, as well as an R' in lead V1 of 4 mm.

nosis could be arrived at only at operation. In Cases 1 and 2, the combination of an atrial septal defect, an electrocardiogram with a frontal plane axis towards the left (0°) (Pryor, Woodwark, and Blount, 1959; Burchell, DuShane, and Brandenburg, 1960) (Fig.), murmurs of mitral regurgitation and mitral regurgitation on left ventriculography in Case 1, suggested a diagnosis of an endocardial cushion defect. In Cases 3 and 5, the diagnosis was only made at the time of operation, an isolated ostium secundum type of septal defect alone being suspected beforehand. At the time of operation, the mitral cleft in Cases 2 and 5 was recognized only when a thrill was noted after closure of the septal defect and the patient taken off bypass. Preoperatively, the regurgitation may have been of small degree, and produced little murmur. It is well known that mitral regurgitation will assume greater importance after a coexisting atrial septal defect is closed (Kirklin and Wallace, 1970), and it is obviously of great importance that the mitral regurgitation be recognized before or at operation, so that repair of the cleft valve can be accomplished. A murmur of mitral regurgitation has appeared postoperatively in patients having closure of an ostium

secundum atrial septal defect (Pocock and Barlow, 1971). In these cases it was felt that this was due to the development of the billowing posterior mitral leaflet syndrome. In our cases, if the thrill of mitral regurgitation had not been detected at operation, the genesis of the murmur found postoperatively would have been obscure and troublesome. It is interesting that our first patient had not only a cleft in the anterior leaflet of the mitral valve, but also a prolapsing, cleft posterior leaflet.

The P wave of the electrocardiogram in Cases 1, 2, 4, and 5 showed left atrial enlargement. While this has been reported to occur in a minority of cases of uncomplicated atrial septal defect, the P wave in this disorder usually suggests right atrial disease. Therefore, the finding of left atrial enlargement in 4 of our 5 patients, especially as manifested in lead V1, pointed to concomitant mitral valve disease, as has been suggested in ostium primum atrial septal defects (Sánchez-Cascos and Deuchar, 1963). The chest x-rays were of no help in this regard. It is possible that echocardiography would have detected an enlarged left atrium preoperatively (Hirata *et al.*, 1969).

The moderate to large size of the atrial septal

defects in our 4 patients with ostium secundum lesions was suggested at the time of catheterization by the small differences in the pressure tracings from the left and right atria (Table 1). Therefore, the atria functioned as a large, compliant chamber, helping to explain the lack of a prominent 'V' wave in 4 of these patients with mitral regurgitation. This has also been found in patients with ostium primum atrial septal defects and cleft mitral valves (Evans, Rowe, and Keith, 1961). Only in Case 4 was a large 'V' wave present in the wedge tracing, and at operation a smaller sinus venosus defect was present in association with more severe mitral regurgitation. The lack of a prominent 'V' wave, then, in the wedge or left atrial pressure tracing of a patient with an atrial septal defect does not rule out mitral regurgitation. Only a left ventriculogram will give a definite preoperative assessment of valvular competence, and in addition will allow the separation of patients with endocardial cushion defects from those with secundum type atrial septal defect and mitral valve cleft (Baron *et al.*, 1964).

The position of the mitral valve cleft was of great interest in our patients. Two had typical clefts in the anterior leaflet, and it could be argued that this represented the chance association of two congenital defects, the common atrial septal defect (Dave *et al.*, 1973), and the uncommon isolated cleft of the anterior leaflet of the mitral valve, possibly representing a form of endocardial cushion defect (Edwards *et al.*, 1965). Indeed, ostium secundum atrial septal defects have been reported to occur in association with the more common partial form of endocardial cushion defect (Evans *et al.*, 1961; Rogers and Edwards, 1948). The origin of the cleft in the posterior leaflet in Case 1 (who also had a cleft anterior leaflet and prolapsing posterior leaflet), and Cases 2, 4, and 5 is more obscure. These clefts were located in the mid-portion of the posterior leaflet and tended to be small in extent. No cleft extended to the mitral annulus, and in Case 1 the cleft was felt to be small enough to require no repair. They were most probably related to the normal tripartite structure of the posterior leaflet (Lam *et al.*, 1970; Ranganathan *et al.*, 1970). The clefts were of some haemodynamic significance, as judged by the thrill noted over the posterior left atrium at the time of operation. In addition, the mitral regurgitation produced by these clefts, had they not been corrected at operation, may very well have led to diagnostic difficulties on their discovery postoperatively.

The importance of recognizing the existence of a mitral valve cleft in association with a secundum atrial septal defect is further shown by the clinical course of Case 4. In this patient, bacterial endo-

carditis developed in what seemed like a straightforward case of secundum atrial septal defect. While endocarditis has been reported to occur with isolated secundum defects, it is extremely unusual (Gault *et al.*, 1968; Vogler and Dorney, 1962; Cohn, Morrow, and Braunwald, 1967; Zaver and Nadas, 1965; Danilowicz, Reed, and Silver, 1971). This is also true in sinus venosus atrial septal defects (Davia, Cheitlin, and Bedynek, 1973). More commonly, endocarditis occurs with ostium primum defect associated with mitral valve cleft (Rogers and Edwards, 1948) or with ostium secundum defect associated with the syndrome of posterior mitral leaflet prolapse (Pocock and Barlow, 1971; Danilowicz *et al.*, 1971). When the diagnosis of mitral valve pathology is made, therefore, bacterial endocarditis prophylaxis should be instituted.

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Requests for reprints to Dr. E. W. Hancock, Cardiology Division, Stanford University School of Medicine, Stanford, California 94305, U.S.A.